

Looking for Answers

Providing Research, Education and Support since 1988

In ASAP's continual efforts to find answers its Board of Directors voted to fund three new research initiatives.

Proposed study from Duke

Given the consistency in evidence for association across two independent studies (Spanish and US) in the ALDH1A2, CDX1 and FLT1 genes, Duke proposes a targeted deep sequencing study of these three genes to identify potentially causal variants. This approach is similar to how they previously identified mutations in the GDF6 and GDF3 genes in CMI families (Markunas et al, 2013).

Washington University

Principal Investigator David Limbrick, MD, PhD will look at the 'Effect of Demographic and Socioeconomic Factors in the Diagnosis and Treatment of Chiari I Malformation'. By analyzing these effects, this study hopes to understand and ultimately reduce disparities in the diagnosis and treatment of CM+/-SM.

Stanford Medical

Principal Investigator Jennifer Quon, MD will conduct a study to establish "quality indicators" for the surgical management of Chiari I malformation. Using an evidence-based approach to establish Chiari quality indicators to inform clinical and surgical decision-making, this study proposes to identify disease-specific instruments that could be implemented in the ambulatory care setting and prospectively validates these metrics to ensure they accurately reflect the range of outcomes seen in this disease. The success of this innovative project will be measured by the 1) seamless integration of quality metrics into our daily workflow; 2) improve patient care and outcomes; and 3) lead to a national collaborative multi-institutional effort focusing on quality outcome metrics for Chiari I malformation.



Syringomyelia Awareness

Thirty years ago, Barbara White decided to go against the advice of family and friends and create an organization that would help others like herself who were diagnosed with syringomyelia. After diligent research, she was able to incorporate in May 1988.

For several years we have been encouraging our members to create a syringomyelia awareness event each May. The number of state proclamations increase each year. We have created new fundraising events (Syringo-My-What Steps Across the States and Kicks & Licks) to promote awareness as well as raise funds for programs and research. Funds raised can also earn members LEARN points to attend the annual conference.

Raising awareness doesn't have to be complicated or consume a lot of time and energy. It can be as simple as sharing information. Don't forget to Tweet #maysyringomyeliaawarenessmonth. Share information about the disorder on social media. Create a Pinterest board. Contact your local newspaper about doing a story in the Lifestyle or Health section. Create a YouTube video about your journey.

Anyone interested in holding a fundraiser can contact Patrice Schaublin for additional information. She will provide suggestions and a detailed package.. Patrice_Schaublin@ASAP.org

"Be strong, you never know who you are inspiring."

Start making plans now to attend the ASAP Chiari & Syringomyelia Conference July 19-23, 2017 Long Island, NY

ASAP Conference

The 2017 ASAP Chiari & Syringomyelia Conference will take place at the Long Island Marriott in Uniondale, New York. Patrice Schaublin negotiated an unbelievable room rate of \$135 (plus tax) per night for conference attendees. You can make reservations by calling the hotel directly at 516-794-3800 or Marriott reservations 1-800-228-9290.

The meeting will start with our opening reception on Wednesday evening, July 19 and run through mid day on Sunday, July 23. Planning is in the early stages but we will release an agenda as soon as it becomes available.

2016 Conference Videos Available Online

You can now watch presentation from the ASAP Chiari and Syringomyelia Conference that took place in San Francisco. Over 30 speakers talked on a variety of subjects pertinent to the Chiari and syringomyelia community.

The videos are available on our YouTube channel and website. Due to the long IP addresses the easiest way to explain is; go to www.ASAP.org. For YouTube click on the icon in the upper right section of any page. To watch on our website, look for the rotating text under the picture section of the home page. Click on the 'Watch Educational Videos' message and follow the leads.

Correction

In the Summer 2016 issue of *Connections*, the ASAP Telephone Outreach incorrectly stated the call takes place the third Thursday each month. **The teleconference call takes place the second Thursday of each month at 7:30 PM Eastern Time.** Please contact the ASAP office by calling 1-800-272-7282 and request an outreach brochure if you are interested in participating. Please leave your name and complete postal mailing address and state that you would like a copy of the Outreach brochure.

Welcome Bridget Borys



Bridget Borys was elected to the ASAP Board of Directors during the October meeting. She graduated from SUNY Fredonia in 2007 with her Bachelors in Social Work. Currently employed by Ontario ARC as a Medicaid Service Coordinator, Bridget enjoys helping those with developmental disabilities.

Bridget lives in Seneca Falls, NY with her husband Michael 'Yogi'.

Yogi was diagnosed with Chiari and syringomyelia in 2001 when running wind sprints training at the police academy. Thankfully a doctor at the local emergency room knew about Chiari malformation and syringomyelia and ordered the MRI.

In 2007 Bridget and Yogi held their first fundraiser with a friend's band to raise money for ASAP programs. Since that time she has been raising funds and spreading awareness for both Chiari malformation and syringomyelia. Bridget has helped organize several walks in the past few years. Most recently, with the assistance of her family, she created a Community Fun Day to spread awareness of Chiari malformation, syringomyelia, ASAP, our mission and vision.

Bridget enjoys helping others and tries to see the positive in everything she does. She looks forward to working with others on this amazing journey with ASAP.

Please Note: Articles in this newsletter are not intended as a substitute for medical advice and do not necessarily represent the viewpoints of the editor, Medical Advisory Board or Board of Directors. Please contact your doctor before engaging in any new therapy or medication.

A Word From Our President



Happy New Year to Everyone!

Whether it be a stranger, a friend, a loved one or a lost connection, offer them a smile and a "Happy New Year," and it will almost surely be returned! If you haven't tried it, please do, it will brighten the day for both of you!

ASAP enters this new year filled with seeking new opportunities and setting new goals. In doing so, it's important to remember last year as well. Our staff, board members, medical advisory board, and volunteers have put in endless hours. At ASAP when one event ends we are onto the next, whether it be a walk, support group, fundraiser, national conference, new awareness campaign, etc. That's the way we always do it! Why? So that we may continue to serve the CM/SM community. Thank you for giving us a wonderful 29 years.

ASAP is constantly striving to serve you, bringing new programs and research to the forefront. In this past year we accepted three new research grants, started the first event to include our canine friends affected by CM/SM, added new board members and new medical advisory board members. We brought on new volunteers, held our 28th annual national conference and awareness events. It is our intention to continue this legacy in the new year. We will continue to fulfill our mission statement, "to improve the lives of persons affected by syringomyelia, Chiari malformation and related disorders while we find the cure."

My passion over the last 20 years has been to serve the ASAP community, spread awareness of Chiari and syringomyelia, and help those affected. I am very fortunate to serve all of you as president of ASAP. It is through this service that I find deep gratification and hope that I have, in some small way, improved lives. There is no better feeling.

Best to all in 2017!

Gentle Hugs always,
Patrice
Patrice_Schaublin@ASAP.org

www.ASAP.org

2017 Conference Co-Hosts



Paolo Bolognese, MD is the director of the Chiari Neurosurgical Center NSPC in Lake Success, New York. A native of Torino, Italy, he graduated magna cum laude in 1986 from the Medical School of the University of Turin. In 1990, he completed his neurosurgical training at the same university under the guidance of Professor Victor A Fasano, an

international leader in the field of high-tech applied to neurosurgery.

During this time, Dr Bolognese became the leading worldwide expert in the field of laser Doppler flowmetry applied to neurosurgery and the top European figure in the field of neurosurgical intraoperative ultrasound.

In 1992 he accepted the invitation of Dr Thomas H Milhorat to transfer his laser Doppler research to the United States and to be retrained under Dr Milhorat at SUNY Health Science Center at Brooklyn. At SUNY at Brooklyn Dr Bolognese completed his residency in neurosurgery, his fellowship in management of Chiari I malformation and related disorders, and served as chief resident in neurosurgery.

In August 2001, Dr Bolognese joined Dr Milhorat at the Department of Neurosurgery at North Shore University Hospital and Long Island Jewish Medical Center as an Associate Director of the Chiari Center where he served until 2014.

In September 2014, he joined Neurological Surgery, PC, as the Director of The Chiari Neurosurgical Center at NSPC.

Dr Bolognese is planning to bring speakers from around the world to speak at this year's meeting. He has already received confirmation for several. In addition to our Medical Advisory Board there will also be some new faces from specialists in the US. He is a proponent of the patient and is determined to provide a conference that will not only educate the patient but the medical community.



Roger W Kula, MD, FAAN completed medical school in 1970 at The Johns Hopkins University School of Medicine in Baltimore, Maryland. He began his internal medicine training at The New York Hospital-Cornell Medical Center. His neurology training continued at the University of California Hospitals, San Francisco, where his exposure

to the influence of then-chairman Robert A Fishman, MD, first stimulated his interest in spinal fluid physiology. He completed his formal residency training at the Medical Neurology Branch of the National Institute of Neurological Diseases and Stroke, Bethesda, Maryland, in 1975, where he continued training in neuromuscular diseases under the mentorship of W King Engel, MD.

In 1977, he returned to New York to establish a neuromuscular disease program at the SUNY Health Science Center at Brooklyn as assistant professor of neurology. He was certified by the American Board of Internal Medicine in 1975 and by the American Board of Psychiatry and Neurology in 1977. He went on to establish one of the most clinically active MDA clinics in the tri-state area and developed a national reputation in the study and treatment of autoimmune neuromuscular diseases, motor neuron diseases and muscular dystrophy.

In 1991, he was appointed chairman of neurology at The Long Island College Hospital while continuing to serve as associate professor of clinical neurology and vice chairman of the Department of Neurology at the SUNY Health Science Center at Brooklyn. He is listed in Who's Who in America, Who's Who In Science and Engineering, New York magazine's "Best Doctors in New York" and Castle Connolly's Best Doctors In America.

In 2003, he joined the Chiari Institute as its Medical Director. Since 2003, Dr Kula has directed the Neuromuscular Clinics at Cohen Children's and LIJ. In 2016 he joined Dr Bolognese at the Chiari Neurosurgical Center NSPC.

Chronic Pain

The annual cost of chronic pain in the United States is as high as \$635 billion a year, which is more than the yearly costs for cancer, heart disease and diabetes according to a study by the American Pain Society. Currently, opioids are the primary treatment option for moderate-to-severe chronic and acute pain. However, innovative technologies like the FDA-cleared Sprint Percutaneous Peripheral Nerve Stimulation (PNS) system from SPR Therapeutics, are offering physicians important drug-free treatment alternatives.

Neuropathic Pain

Neuropathy occurs when nerves are injured from trauma or disease and can also be the result of a surgical procedure. This type of pain, unlike physiological pain, persists even after the injured nerve has healed and is often resistant to pain relievers like acetaminophen and naproxen. While opiates are used to alleviate pain, they have side effects and are not always effective for neuropathic pain patients.

Could My Symptoms Be Something Else?

You may feel like your doctor(s) is not validating your disorder when they suggest testing for something else. The truth is, many other disorders might be easier to treat, and multiple disorders do have the same symptoms. We all want to feel better so be open to your doctor's suggestions.

Could your symptoms be anemia?

Some common symptoms of anemia may include:

- Weakness
- Dizziness
- Pale skin
- Headache
- Numbness or coldness in hands and feet
- Low body temperature

Anemia develops when you don't have enough robust, healthy red blood cells to carry oxygen throughout your body. People with anemia have less oxygen in their blood, which means the heart must work harder to pump enough oxygen to their organs. Cardiac-related symptoms include arrhythmia (an abnormal heart rhythm), shortness of breath, and chest pain.

About one in seven children develop anemia by age 2, most often because they don't have enough iron in their diet. People who have iron-deficiency anemia may feel the urge to eat inappropriate things like dirt, clay, ice, or starch, a behavior called pica. Pediatricians test all children for anemia at 12 months. Without treatment, a severe case of anemia could permanently affect brain development.

Women and people with chronic diseases have the greatest risk of anemia. Chronic diseases such as kidney disease can affect the body's ability to make red blood cells. A diet low in iron, folate, or vitamin B12 also increases your risk. And some types of anemia are hereditary.

A diet that's low in iron can cause anemia. Iron from plants and supplements isn't absorbed as well as the iron in red meat. Digestive concerns such as Crohn's disease, celiac disease, or even having gastric bypass surgery can interfere with iron absorption. And some foods and medicines can hinder iron uptake when taken with iron-rich foods. They include:

- Dairy
- Other calcium-rich foods
- Calcium supplements
- Antacids
- Coffee
- Tea

The body needs both vitamin B12 and folate to make red blood cells. A diet too low in these vitamins sometimes can cause anemia. An autoimmune disorder or digestive problem also can prevent your body from absorbing enough B12. Animal-based foods and fortified breakfast cereals are good sources of B-12. Folate is in leafy green vegetables, fruits, dried beans, and peas, and is added to breads, pastas, and

cereals as folic acid.

Chronic illness or infection can cause the body to make fewer red blood cells. This can result in a mild drop in hemoglobin. If you have significant blood loss, then you may develop iron-deficiency anemia. And some drugs and medical treatments can also put you at risk for anemia. Consult your doctor to see if you need iron or other supplements.



A complete blood count test will check your levels of red blood cells, white blood cells, platelets, and hemoglobin. It will also check other factors such as average size, variability in size, volume, and hemoglobin concentration of red blood cells. If you have iron-deficiency anemia, your red blood cells may be smaller than normal. Your health care provider also may ask about your symptoms, medicines you take, and your family history.

Iron pills are often needed for anemia that's caused by a deficiency in that mineral. Ferrous iron is more easily absorbed than ferric iron. It's best taken with food, especially orange juice and other foods rich in vitamin C. But don't mix your iron pill with calcium, coffee, or tea, which can block absorption. And never take iron without a doctor's order or let children near the pills. An iron overdose can be dangerous. Some people may need folic acid or vitamin B12 supplements, too. You can prevent some types of anemia with a healthy diet.

Foods containing iron include lean red meat, liver, fish, tofu, lentils and beans, dark green leafy vegetables, and dried fruits. Also eat foods with vitamin B12 and folic acid, such as eggs and dairy products, spinach, and bananas. Many breads, cereals, and other foods are fortified with all three key nutrients: iron, B12, and folic acid. Vitamin C found in citrus and other fruits as well as vegetables will help your body absorb iron.

Too much iron can cause serious problems. Iron overload can be a result of repeat blood transfusions or an inherited condition, but taking too much iron also is a risk. The many symptoms of iron overload are related to excess iron depositing into organs and causing problems in the liver, heart, and pancreas. Iron levels can be reduced through phlebotomy (blood removal) or medications.

Treating your anemia and eating a well-rounded diet can give you more energy and enhance your life. Most people can manage their anemia through a healthy diet and iron or vitamin supplements, if a doctor says they are deficient in one of the key nutrients. If you have a chronic disease, then good management of your condition also will help you prevent or manage anemia.

**Molly's 2nd Annual
Chiari & Syringomyelia Walk and Roll**



**Oklahoma City ASAP Syringomyelia and
Chiari Walk & Roll for a Cure**



Thanks Molly and family for another GREAT event!



September 17, 2016
Germantown, MD



Special thanks to Ann Humphreys and the OKC Support Group

September 24, 2016
Oklahoma City, OK



Remember ASAP..

When It's Time to Remember Loved Ones

Our appreciation to everyone who made a recent donation to ASAP on behalf of their friends and loved ones.

We will send an acknowledgement card to individuals or families when you make a \$5 (or more) donation to the organization. Please indicate whether the gift is 'in honor of' or 'in memory of' and provide name and mailing address of the recipient.

In Honor of Person

- Donor
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Donalda Wolfe
- Austin Rodriguez**
Teri's Run & Twilight Walk
- Jeni Coonrad Adair**
Cynthia Jankowski
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Jeanne Swack



ASAP History



The first ASAP newsletter was written by Barbara White and consisted of 4 pages of typed information. Barbara continued to send out letters to the members

for several years. During that time, she underwent multiple unsuccessful surgeries that eventually left her a quadriplegic.

In 1990, ASAP Board member Candace Morse volunteered to take over the responsibilities of the newsletter. In an age before social media and the internet, the newsletter was the preeminent method of letting members know what advances the organization was making in its journey for knowledge and awareness.

In October 1991, Staci Wietrecki created the first KiDS page. She was fourteen at the time and a veteran of the Chiari /syringomyelia community. Over the next few years, Staci's endeavors on behalf of those who share her battle would result in her being awarded the 811th Point of Light from President George H W Bush.

In 1993 the newsletter got a new name *Syringomyelia Connections* when volunteer Angela Smith took over as editor. The name was the result of a contest Angela held to 'name the newsletter'. Angela continued as editor until 1996 when Christian Barthol became the new editor. Like Barbara, Christian was a quadriplegic. He continued as editor until 2001.

Contact Information

American Syringomyelia & Chiari Alliance Project

Mailing Address:
PO Box 1586
Longview TX 75606-1586

Physical Address:
300 North Green Street, Suite 412, Longview, Texas 75601

Phone: 903-236-7079

Fax: 903-757-7456

Toll-free: 800-ASAP-282

Email: info@ASAP.org

Staff:

Patricia_Maxwell@ASAP.org

Jamie_Mayhan@ASAP.org

Autonomic Dysreflexia

The autonomic nervous system controls involuntary actions such as blood pressure, heartbeat, and bladder and bowel function. Autonomic dysreflexia is a life-threatening reflex action that primarily affects those with injuries to the neck or upper back. It happens when there is an irritation, pain, or stimulus to the nervous system below the level of injury. The irritated area tries to send a sensory signal to the brain, but the signal may be misdirected, causing a runaway reflex action in the spinal cord that has been disconnected from the brain's regulation. Unlike spasms that affect muscles, autonomic dysreflexia affects blood vessels and organ systems controlled by the sympathetic nervous system. Anything that causes pain or irritation can set off autonomic dysreflexia, including a full bladder, constipation, cuts, burns, bruises, sunburn, pressure of any kind on the body, or tight clothing. Symptoms of its onset may include flushing or sweating, a pounding headache, anxiety, sudden increase in blood pressure, vision changes, or goose bumps on the arms and legs. Emptying the bladder or bowels and removing or loosening tight clothing are just a few of the possibilities that should be tried to relieve whatever is causing the irritation. If possible, the person should be kept in a sitting position, rather than lying flat, to keep blood flowing to the lower extremities and help reduce blood pressure.

Signs and Symptoms

The symptoms of autonomic dysreflexia may include:

- Anxiety and apprehension
- Irregular or racing heartbeat
- Nasal congestion
- High blood pressure with systolic readings often over 200 mm Hg
- A pounding headache
- Flushing of the skin
- Profuse sweating, particularly on the forehead
- Light-headedness
- Dizziness
- Confusion
- Dilated pupils

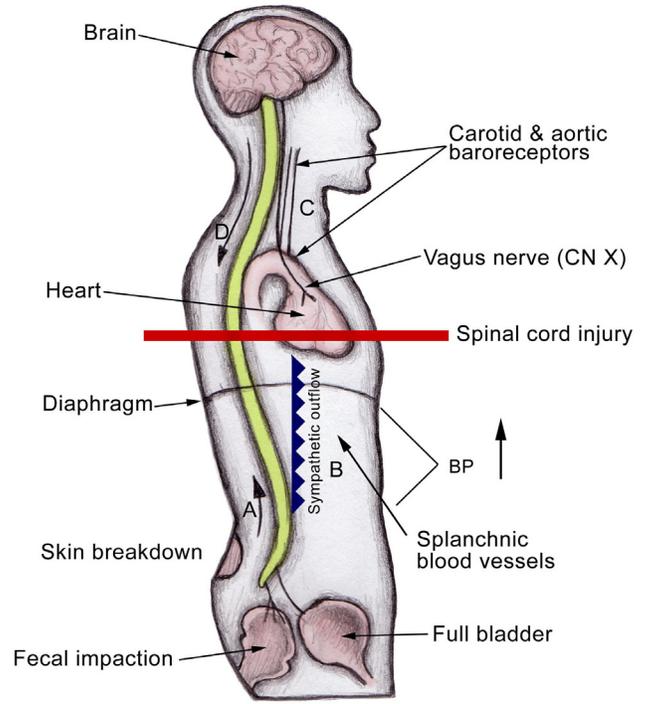
What to do when autonomic dysreflexia is triggered

If autonomic dysreflexia is suspected, the first thing to do is sit up or raise the head to 90 degrees. If you can lower your legs, do so. Next, loosen or remove any constricting clothing, and be sure to check your blood pressure every five minutes.

An individual with spinal cord injury above T6 often has a normal systolic blood pressure in the 90-110 mm Hg range.

A blood pressure reading of 20mm to 40mm Hg above baseline in adults may be a sign of autonomic dysreflexia. 15mm above baseline in children, and 15mm to 20mm above baseline in adolescents may be a sign of autonomic dysreflexia .

Most importantly, locate and remove the offending stimulus, if possible. Begin by looking for your most common causes: bladder, bowel, tight clothing, skin issues. Keep in mind as you remove the cause that your autonomic dysreflexia may get worse before it gets better.



- A strong sensory input (not necessarily noxious) is carried into the spinal cord via intact peripheral nerves. The most common origins are bladder and bowel.
- This strong sensory input travels up the spinal cord and evokes a massive reflex sympathetic surge from the thoracolumbar sympathetic nerves, causing widespread vasoconstriction, most significantly in the subdiaphragmatic (or splanchnic) vasculature. Thus, peripheral arterial hypertension occurs.
- The brain detects this hypertensive crisis through intact baroreceptors in the neck delivered to the brain through cranial nerves IX and X.
- The brain attempts two maneuvers to halt the progression of this hypertensive crisis. First, the brain attempts to shut down the sympathetic surge by sending descending inhibitory impulses. These impulses are unable to travel to most sympathetic outflow levels because of the spinal cord injury at T6 or above. Inhibitory impulses are blocked in the injured spinal cord. In the second maneuver, the brain attempts to bring down peripheral blood pressure by slowing the heart rate through an intact vagus (parasympathetic) nerve; however, this compensatory bradycardia is inadequate and hypertension continues. In summary, the sympathetics prevail below the level of neurologic injury, and the parasympathetic nerves prevail above the level of injury. Once the inciting stimulus is removed, reflex hypertension resolves.



ASAP's Mission: to improve the lives of persons affected by syringomyelia, Chiari malformation and related disorders while we find the cure.

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ASAP Connections

The American Syringomyelia & Chiari Alliance Project (ASAP) is a tax-exempt 501(c)(3) organization. Our goals include providing a clearinghouse for information on syringomyelia (SM), Chiari malformation (CM), and related conditions.

We offer a supportive network of programs and services and fund research to find better therapies and cures. ASAP is supported by tax deductible donations.

ASAP Connections is published quarterly for ASAP members. Your contributions of articles, letters, and photos are encouraged. The editor reserves the right to edit any article in order to accommodate space.

Email: Patricia_Maxwell@ASAP.org

Mail: *ASAP Connections*
PO Box 1586
Longview TX 75606

ASAP Connections Editor: Patricia Maxwell